ESKRIDGE (J.S.)

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IDIOPATHIC MUSCULAR ATROPHY COMPLICATED BY MULTIPLE NEURITIS.[1]

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THE case to which I wish to call your attention today is that of Miss A. R., aet. 46, Michigan, in Coloradonine years. She gives her family history as follows: Father died of slow paralysis, which finally affected speech and respiration. Knows of no other case of nervous disease in father's family. No nervous disease in mother's family. One brother, aged about thirty-five years, a tinner by occupation, has been a resident of the hospital for some time and is affected with progressive muscular atrophy of a peculiar type. One sister, aged about forty-nine, is living and apparently well. In the brother's case the paralysis and muscular atrophy affect both the upper and lower extremities, but mainly the upper. The patient was well up to her seventh year, when she was scalded by sitting down in a pail of boiling water. The parts were severely burned, and the accident seemed to produce considerable shock. She thinks she was not as well afterwards. She suffered from the ordinary diseases incident to childhood, but enjoyed fair health up to her seventeenth year, when she first noticed a weakness in the muscles of the legs, which she expresses as weakness of the knees. So far as she knows there was no apparent cause for this. The weakness was not accompanied with pain. During the next six years, the disease did not seem to make much progress, and does not seem

[1] A clinical lecture delivered at the Arapahoe County Hospital, Oct. 8, 1892. Stenographically reported by Miss Lottie M. Page.

to have been attended by much, if any, muscular wasting; and she was only inconvenienced by a tendency of the legs to give way occasionally after standing a long time. The arms as yet were not affected. In her twenty-third year, her legs suddenly refused to support her while she was going down stairs, and she fell a distance of about five feet, striking on her spine in the mid-dorsal region. She experienced pains in the small of the back, and some pain in the right shoulder. The latter, she was told, was injured. After this accident she was much more uncertain in her gait, so that she found it very difficult to walk. The muscles of the legs since have become quite weak and wasted. She suffered with little or no pain in the legs until about eight years ago, when a dull, aching, uneasy, nervous pain began. She has suffered more or less with sharp, shooting pains in the small of the back when trying to walk, or while being on her feet. She makes no complaint of the hands or arms.

Condition, November 15th, 1891: She is unable to stand or walk without assistance. There is marked foot-drop. She is unable to dorsally flex either foot. Plantar flexion very slight, an attempt to plantar-flex the right foot causes a slight dorsal flexion of the great toe The muscles of the legs below the knees are wasted. and flabby. She is unable to extend either leg at the knee. All muscles below the knees are paralyzed except a few fibres of the posterior tibial group. Flexors and extensors of the thighs are decidedly paretic, the extensors more so than the flexors, and this is more marked in the right than the left leg. Reflexes: Plantar: R., present only for the greater and adjacent toe; L., irritating plantar surface causes no movement of the toes, but slight inversion of the foot takes place. Ankle clonus, absent. Knee-jerks, absent. Inguinal reflexes, present. Epigastric reflexes present. Dynamometer, neither hand is able to move the indicator. No wrist drop. Extensors and flexors of the wrist, and biceps and triceps on both sides still retain slight power; greater on the right side. All reflexes in the arms absent. Tongue protrudes in median

line. No facial paralysis. Pupils normal in size, equal, and respond to light and accomodation. No paresis or paralysis of any of the ocular muscles. Measurements: Calf, R., 9 ½ inches; L., 9 ¼. Thighs, R., 14 inches; L., 13 ¾. Fore-arms lax, R., 6 ¾; L., 7. In contraction, R., 7; L., 7 ¼. Biceps, R., 9 inches; L., 9 ¼. In contraction, R., 9 ⅝; L., 9 $\frac{7}{16}$. Both deltoids fairly strong and but little wasted. The great pectoral muscles, inferior and superior fibres, are paralyzed for each of their separate actions, but superior fibres contract feebly in bringing arms across the chest, while the inferior, or costal, fibres, remain inactive. The pectoral muscles on both sides are greatly wasted. Each thenar eminence is considerably wasted. Hypothenar eminences nearly normal in size. No distinct wasting of any other groups of muscles of the hands, although the hands seem unnaturally small for a woman of her size. Right trapezius and serratus magnus muscles very weak and greatly wasted; while these muscles on the left side are fairly strong, and but slightly wasted.

Reactions to the faradic current: The anterior tibial and peronei muscles on both sides fail to respond to the strongest current. The posterior tibial group on each side respond feebly to a strong current. There is some lessening of muscular irritability to the faradic current in the muscles of the thighs. The muscles of the hands respond feebly to a strong current. Those of the arms and shoulders respond better, but not normally. As no galvanic battery is at hand, the muscles can not be tested by this current.

There is slight tenderness over the spines of the first and second lumbar vertebræ. She complains considerably of pain in the back and posterior portion of the chest, but of none in legs or arms. No tenderness over the nerves of legs or arms.

Tactile sense is absent in each leg, from a point just above the knees down, including the feet, but is present and apparently normal in all other portions of the body.

Pain sense is present throughout the body, and seems

acute in the anæsthetic areas, but normal in other portions of the body. Posture sense in feet is absent, but present in legs and arms. Localization sense is perverted in legs below the knees. A pin prick in one leg is spoken of as occurring in the other leg, at a point corresponding to the point irritated, a condition known as allocheiria. Pressure sense lessened in legs below the knees. All forms of sensation normal in hands, arms and trunk. Vision, hearing and taste normal. Smell absent.

October 8th, 1892. We will re-examine the patient to-day, and compare her condition with the results of the examination made nearly a year ago.

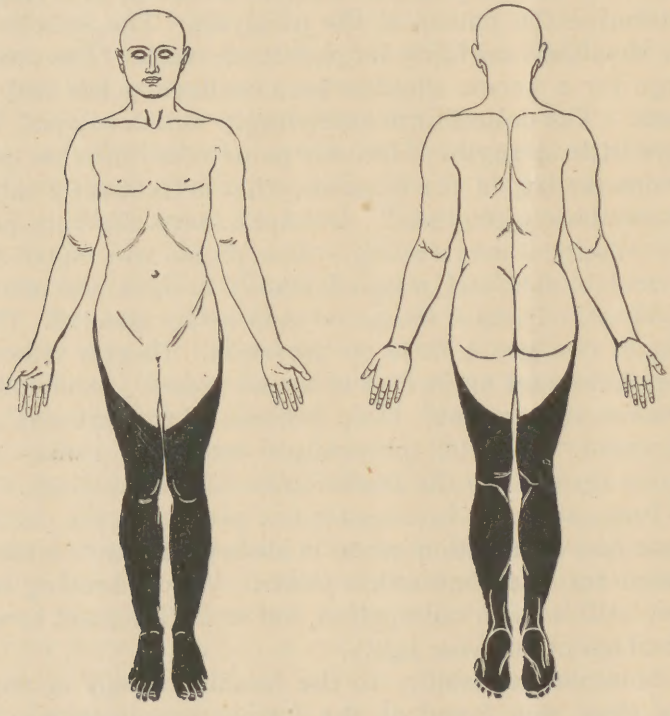
There is marked foot-drop, with inversion of both feet. All the muscles below the knees are completely paralyzed and greatly wasted. She is unable to extend or flex either leg at the knee. When she endeavors to extend the legs at the knees there is perceived a feeble contraction of a few muscular fibres, but it is too weak to move the leg. The flexors of the knees are a little stronger than the extensors, but their strength is insufficient to move the legs more than a few inches even when the feet are lying on the bed. The flexors and extensors of the thighs are exceedingly weak, but they are stronger than the thigh muscles that should move the legs at the knees. The muscular wasting is considerable in the thighs, but not so great as in the legs. The hip muscles are wasted, but to a less extent than those of the thighs. She has not been able to retain urine well during the last two or three years,especially in coughing. There is no paralysis of the sphincter of the bowels. It is probable that the loss of control of the bladder is due more to prolapsus of the womb, from which she suffers, than to any paralysis of the sphincter of the bladder. There seems to be no dribbling of urine nor involuntary evacuation of the contents of the bladder, except during fits of coughing. The trunk muscles are not completely paralyzed, but they are weak, and she is unable to sit unless she receives some support. Extensors of each wrist are stronger than flexors, but both are weak. There is absolute paralysis

and considerable wasting of the muscles of the hands. Dynamometer, R., O; L., O. The biceps are fairly strong the triceps are much weaker. The deltoid muscles are not completely paralyzed but nearly so. Yet you observe that they seem abnormally developed. The pectoral muscles are very weak, and greatly wasted. The wasting in upper arm, with the exception of the pectorals is mainly below the elbows. I want to call your special attention to this point, as I shall consider it in trying to determine the nature of the paralysis. The muscles of the shoulders are fairly large, but are weak. The arm is large for a person who has been confined to her bed for years. The deltoids are exceedingly well developed, but have little strength. Her only pain in the limbs has been a sore feeling in the muscles; what is frequently called "muscular rheumatism." It is not a sharp, shooting, pain but simply a sore feeling. The pupils are equal and normal in size, and respond readily to light and accommodation. None of the ocular muscles are affected. The tongue deviates a little to the right. Plantar reflexes, knee-jerks and ankle clonus are all absent. Abdominal reflexes are present. Deep reflexes of the fore-arm are increased. Tactile, temperature and pain senses are absent throughout the shaded areas of the drawings.

Pressure sense is lessened, but present in the shaded areas and localization sense is absent. Posture sense is absent for knee and ankle joints. Vision, hearing and taste still remain unimpaired, but smell is absent as was found about one year ago.

Muscular irritability to the faradic current is much less than was found at the former examination. No response to the strongest current can be obtained in muscles below the knees, except in a few fibres that slightly move the great toe of the left foot in the plantar direction. The muscles of the thighs, hips and arms, only feebly respond to a very strong and painful current. During the last year wasting in the muscles of the legs and arms has increased considerably, and it has been most pronounced in the muscles below the knees and

elbows; muscular weakness has become very much more marked, and electrical irritability has greatly lessened; anæsthetia has extended well up the thighs, and loss of pain and temperature senses have become absolute in the anæsthetic areas. From the extreme weakness of the deltoids and some other muscles that still appear but little wasted, it is probable that a pseudo-hypertrophic change is going on in certain muscles.



Let us inquire into the nature of the disease from which our patient is suffering.

A bilateral brain lesion, a chronic myelitis, a spinal pachymeningitis, a polio-myelitis or a multiple neuritis, may give rise to sypmtoms somewhat similar to those presented by the patient before us.

Brain lesions, as a rule, are unilateral, but there are exceptions to this rule of course. There is, in the hospi-

tal, a case, which I will bring before you in a week or two, with a bilateral lesion of the brain, producing paralysis on both sides of the body. When the lesion is in the brain, the muscles of the face are affected and speech is interfered with; if the lesion is vascular in nature it usually comes on more or less suddenly; and it may affect deglutition. These symptoms are all absent; besides, brain lesions do not cause great muscular wasting with flaccidity of the muscles and reactions of degeneration. We can exclude brain lesion as the cause of the symptoms which this woman presents.

Can we exclude chronic myelitis? Chronic myelitis gives rise to paralysis, muscular wasting, loss of faradic irritability of the muscles and anæsthesia, with disturbance of all other sensory phenomena in the parts affected. These are the main symptoms from which the patient before us is suffering, but in chronic myelitis, affecting the cord as high as the cervical region, there are other symptoms which are absent in this case, and the general symptoms are differently associated from what we find them in the patient before us.

In chronic myelitis, extending above the lumbar enlargement of the cord, but involving the lumbar and cervical enlargements, we should have contractures of the muscles of the arms, of the trunk, and possibly to some extent of the legs, although the leg muscles might be flaccid if the lesion in the lumbar enlargement were very severe; the arms and trunk would present sensory disturbances as well as the legs; anæsthesia would be found around the anus, and over the greater portion of the external genitalia, if the feet and legs, below the knees, were anæsthetic, and the sphincters of the bladder and anus would be affected. It is evident, then, that chronic myelitis will not account for the symptoms which we have studied to-day. All the symptoms of chronic myelitis are present in a modified form in chronic spinal pachymeningitis when it is sufficiently severe to give rise to such absolute paralysis as we find in the patient before us; so that we may also exclude this disease.

Is the disease one of chronic polio-myelitis (spinal progressive muscular atrophy)? In favor of this we have gradual wasting and weakness of the voluntary muscles, beginning in the legs, and later involving the arms and trunk muscles, but against chronic polio-myelitis we find marked anæsthesia, and other sensory disturbances in the legs; besides we find here heredity well marked, not of a neurotic taint, but of a characteristic form of progressive muscular wasting. The father died of some disease attended with great muscular wasting, and a brother is suffering from paralysis of the arms, in character, very much like that presented by his sister, save that her paralysis first manifested itself in the legs and is now attended with sensory disturbances. The tendency to a family disease, whose characteristic is progressive muscular atrophy, will justify the diagnosis of idiopathic muscular atrophy. But in this disease there are no marked sensory disturbances, such as anæsthesia, analgæsia, etc. How then can we account for the profound sensory affection presented by our patient? I believe the sensory disturbance is due to a distinct disease, chronic multiple neuritis, limited to the lower extremeties.

It seems to me that we are justified in making the diagnosis of idiopathic muscular atrophy, associated with chronic multiple neuritis, the latter limited to the inferior extremities.

Idiopathic muscular atrophy, (muscular dystrophy), is an extremely infrequent disease, and to have added to it chronic multiple neuritis must be still more infrequent, as this is the first case of the kind that has come under my observation. This form of muscular atrophy is not due to change in the ganglion cells in the anterior horns of the cord, but it is supposed to occur in individuals, usually found in family groups, who have a developmental muscular defect.

The diagnosis of this form of muscular atrophy, from the spinal form, is based largely upon the family tendency of the disease, its beginning in childhood and involvement of the face muscles at times.

The prognosis is not good so far as recovery is concerned, but many cases become arrested in their progress for a number of years, and the more chronic the disease has been in its course the more likely is it to become arrested.

In the way of treatment but little can be done beyond keeping up a moderate amount of exercise for the affected muscles so long as exercise is possible, avoiding over exertion and exhausting influences, stimulating the nutrition of the muscles by massage and electricity, and paying attention to the general health of the patient.